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*Original Article*

# Symptomatic idiopathic pulmonary artery aneurysm: a case report and a mini-review of the literature

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**Abstract:** Pulmonary artery aneurysms (PAAs) are rare, more prevalent in younger population with equal sex incidence. Congenital, idiopathic, autoimmune, infectious, inflammatory, and malignant etiologies have been linked to PAAs. Commonly, patients with PAA are asymptomatic, even those with large PAAs. Presenting symptoms, if any, are non-specific. The management should target the underlying conditions and serial imaging follow-up. Signs and symptoms of disease progression should prompt a change in treatment strategy. Though there is no consensus, those who are symptomatic with a PAA diameter > 5 cm generally should undergo surgical repair. More recently, endovascular interventions are available for certain PAAs. We present a 78-year-old female who was referred to the cardiology clinic for cough and dyspnea. Using computed tomography (CTA) of the chest, she was diagnosed with aneurysm of the main pulmonary artery (PA), without involvement of distal pulmonary arteries or thoracic aorta. She underwent repair of the pulmonary artery using a 34-mm tubular graft with a complete resolution of her symptoms.

**Keywords:** Pulmonary artery aneurysm, aneurysm repair, aneurysmectomy of the pulmonary artery

## Introduction

Pulmonary artery aneurysms (PAAs) are rare abnormalities of the pulmonary vasculature and infrequently diagnosed. Based on 105,571 postmortem examinations at the Mayo Clinic, its incidence is estimated to be 1 in 14,000. PAAs usually occur in the younger population than aortic aneurysms and have equal sex incidence [1].

Aneurysms are defined as a focal dilatation of a blood vessel involving all three layers of the vessel wall while pseudoaneurysms do not involve all three layers of the vessel wall. It is important to distinguish between these diagnoses as pseudoaneurysms have a higher risk for rupture and bleeding [2]. A PAA is described as a focal dilatation of pulmonary artery (PA) beyond maximal normal diameter [3]. There are no standard diameter size parameters to clearly define PAAs [2]. By non-contrast computed tomography (CT), the mean diameter for the main PA in normal healthy adults is 25 mm ± 3 mm with the upper limit of normal in males being 29 mm and the upper limit of normal in females being 27 mm [4]. Generally, PAA can be diagnosed when the main PA diameter is 1.5 times greater than the upper limit of normal (≥ 43 mm in males and ≥ 40 mm in females) [2]. Alternatively, PAA can be diagnosed when the main PA diameter is greater than 45 mm or a branch PA diameter is greater than 30 mm in both males and females [5]. The majority of all PAAs are located in the main pulmonary artery (89%) with the remaining locations within the pulmonary branches (11%) [6]. Additionally, the left PAs were more commonly affected than the right PAs.

The etiologies of PAAs includes congenital, idiopathic, autoimmune, infectious, inflammatory, and malignant cause. Historically, congenital causes were most commonly associated with PAAs and attributed to 50% of the etiology [7]. More recent study [2] showed that congenital causes were only 25% of all cases. In general, the presumed mechanism is that increased flow from a left-to-right shunt leads to increased hemodynamic shear stress on the vessel walls

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allowing aneurysm formation in congenital heart diseases [8]. The most commonly associated congenital heart defects with PAAs are persistent ductus arteriosus, ventricular septal defects, and atrial septal defects [1, 7, 9, 10]. Hypoplastic aortic valve and bicuspid aortic valve have also been identified as significant causes of PAAs [1, 7, 9, 10].

Commonly, patients with PAA are asymptomatic, including those with large PAAs with diameters up to 70 mm [10-12]. Additionally, clinical manifestations are nonspecific which include dyspnea, chest pain, hoarseness, palpitations, and syncope [12-15]. Bronchus compression by a PAA may also cause cyanosis, cough, pneumonia, and fever [10, 12, 16]. Idiopathic PAA may present with hemoptysis and may be a warning for impending rupture [12]. The incidence of pulmonary emboli with PAAs is reportedly high [16]. Rarely, idiopathic PAAs may present with hemothorax or recurrent laryngeal nerve palsy [2]. Other possible associations depending on the underlying etiology include right atrial and ventricular hypertrophy, right heart failure, tricuspid regurgitation due to annular dilatation, and mild pericardial effusion and pleural effusions [13, 14, 16]. PAA dissection is rare but life-threatening and can occur in up to 19% of patients without pulmonary hypertension [12]. About 80% of patients with PAA dissection occur in the main PA trunk and only 15% of these patients are diagnosed alive [10].

PAAs are usually diagnosed incidentally on imaging studies performed for unrelated reasons. PAAs may appear as hilar enlargement, lung nodules, or as a pulmonary mass on a standard CXR prompting additional investigation [8, 10]. The gold standard for diagnosis is pulmonary angiography with the ability to identify the PAA and its involvement with the vascular structures as well as hemodynamic assessment [10]. However, it is invasive and only reveals the interior lumen of the PAA [10, 11, 17]. More common at present, contrast-enhanced CT confirms the diagnosis with the ability to provide additional information such as PAA size, location, and extent [12, 14]. Transthoracic or transesophageal echocardiography is another useful modality to investigate the function of the heart and its valves and possible identification of shunts [10-12, 14].

After a PAA has been diagnosed, the decision for treatment poses another challenge as there are no clear guidelines for the best approach [10]. The management should be tailored to the underlying cause, hemodynamics, and associated comorbidities. A conservative approach comprises medications to target the underlying condition, control of pulmonary hypertension, and imaging follow-up [2]. More invasive surgical options include aneurysmorrhaphy, lobectomy, bilobectomy, aneurysmectomy, and pneumonectomy [2]. Generally, surgical intervention is recommended in symptomatic patients and a PAA with diameter greater than 5 cm [18]. Although, there is no set threshold for intervention, a review by Kreibich et al. [10] suggests surgical repair when the PAA diameter size is greater than 5.5 cm, whereas Seguchi et al. [14] recommends surgical repair when the PAA diameter is greater than 6 cm. More recently, endovascular treatment options have been described [3, 19].

Our case displays an unusual manifestation of a patient with symptomatic PAA. The main objective of this case report is to review the current literature and share our experience of management of this rare clinical entity.

## Case report

A 78-year-old female was referred to the cardiology clinic by her pulmonologist. Her past medical history includes hypertension, gastroesophageal reflux disease (GERD) and irritable bowel syndrome. She had been experiencing persistent cough and intermittent dyspnea for which the work-up was initiated. During her initial work-up, a CTA of the chest was performed revealing an aneurysm of the main pulmonary artery (PA) measuring 5.6 cm in the transverse diameter and the left and right pulmonary arteries were normal in caliber (**Figure 1**). There was no evidence of thoracic aortic aneurysmal dilatation or dissection. There was also no evidence of pulmonary emboli. Pulmonary function testing demonstrated normal spirometry with normal total lung capacity (TLC) and lung diffusing testing (DLCO).

She was hospitalized for uncontrolled hypertension. A CT of the chest without contrast was done showing a PAA measuring 7 × 5.8 cm. Her antihypertensive medications were optimized. Transthoracic echocardiogram showed a normal left ventricular ejection fraction (LVEF) of 60%, mild left ventricular hypertrophy, normal

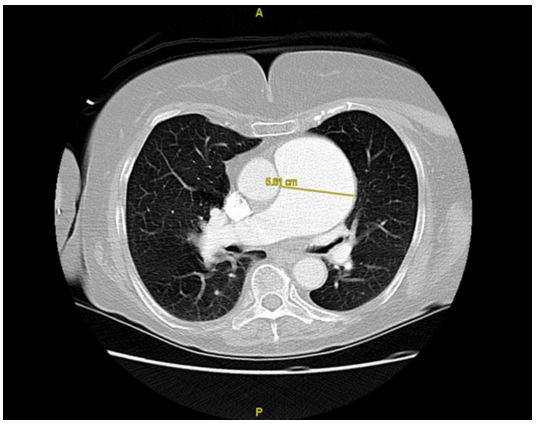


**Figure 1.** Cross section of the CTA chest showing aneurysmal main pulmonary artery measuring up to 5.6 cm in diameter.

right ventricular size and function, normal atrial size bilaterally, and no evidence of significant valvular abnormalities, although the pulmonic valve was not well visualized. There was also no evidence of structural abnormalities such as an atrial septal defect (ASD) or ventricular septal defect (VSD). The pulmonary artery pressure could not be estimated due to an insufficient envelope. She subsequently underwent a transesophageal echocardiogram (TEE) to better assess the pulmonary valve. This study revealed a normal LV systolic function, trileaflet aortic valve without stenosis or regurgitation, mild mitral and tricuspid regurgitation, pulmonic valve with mild to moderate regurgitation and no stenosis. The main PA was dilated measuring 5.8 cm in diameter. The estimated PA systolic pressure was 25-30 mmHg, and diastolic PA pressure was 8-12 mmHg (**Figure 2**). There was no left-to-right shunt. A cardiac catheterization was performed which revealed nonobstructive coronary arteries. She was evaluated by cardiothoracic surgery.

Due to the size of the PAA and her symptoms, she ultimately underwent surgical repair (**Figure 3**). The surgery was performed without cross-clamping the aorta and without cardioplegia. Full cardiopulmonary bypass was placed after adequate anticoagulation was achieved. Intraoperatively, the pulmonic valve was found to be trileaflet and is structurally normal. The main pulmonary artery was resected at the level of the pulmonic valve commissures and is reconstructed using a 34 mm tubular graft by end-to-end anastomosis. The distal main PA was transacted just before its bifurcation and is attached to the graft by end-to-end anastomosis. Intraoperative TEE showed normal functioning right and left ventricle and pulmonic valve without significant regurgitation. She recovered from the surgery without any events with complete resolution of her symptoms.

## Discussion

The clinical course of PAA is insidious and the patients with PAA are generally asymptomatic. PAAs are often diagnosed incidentally. After identification of the PAA, a CTA of the chest should be performed to fully evaluate the size and extend of the PAA including the involvement of the branch pulmonary arteries. Magnetic resonance imaging (MRI) can be used when CT is contraindicated. T1-weighted images are able to detect pseudoaneurysms and another advantage of MRI over CT is the ability to show arterial wall thickening and characterize the direction of blood flow and hemodynamics of the aorta and pulmonary artery [3, 10]. A transthoracic echocardiogram (TTE) should also be performed to assess ventricular function, valvular function and to rule out any intracardiac shunt. Pulmonary valve stenosis has been recognized as an isolated cause of PAAs [8, 10, 11]. Early pulmonary valve commissurotomy may also provoke aneurysmal formation [10, 11]. Many patients with PAA may present with pulmonary valve regurgitation which is likely a result of annular dilatation by the PAA or could be an independent etiology [6, 10, 20]. Frequently, a TEE is performed, which is a better modality to evaluate intra-cardiac shunt, pulmonic valve and pulmonary arteries. If the clinical suspicion for left-to-right shunt is high, and the TEE is not revealing, a dedicated cardiac CT or cardiac MRI should be performed to rule out any left-to-right shunt including extra cardiac shunt, for example, anomalous pulmonary venous return.

Further work up should be directed at identifying any potential underlying etiologies of the PAA. Connective tissue disorders such as Ehlers-Danlos syndrome, Marfan syndrome, and cystic medial necrosis have also been associated with PAAs [15, 21, 22]. Acquired PAAs are another important group to consider which include pulmonary hypertension, autoimmune disease, vasculitis, infection, chronic pul-

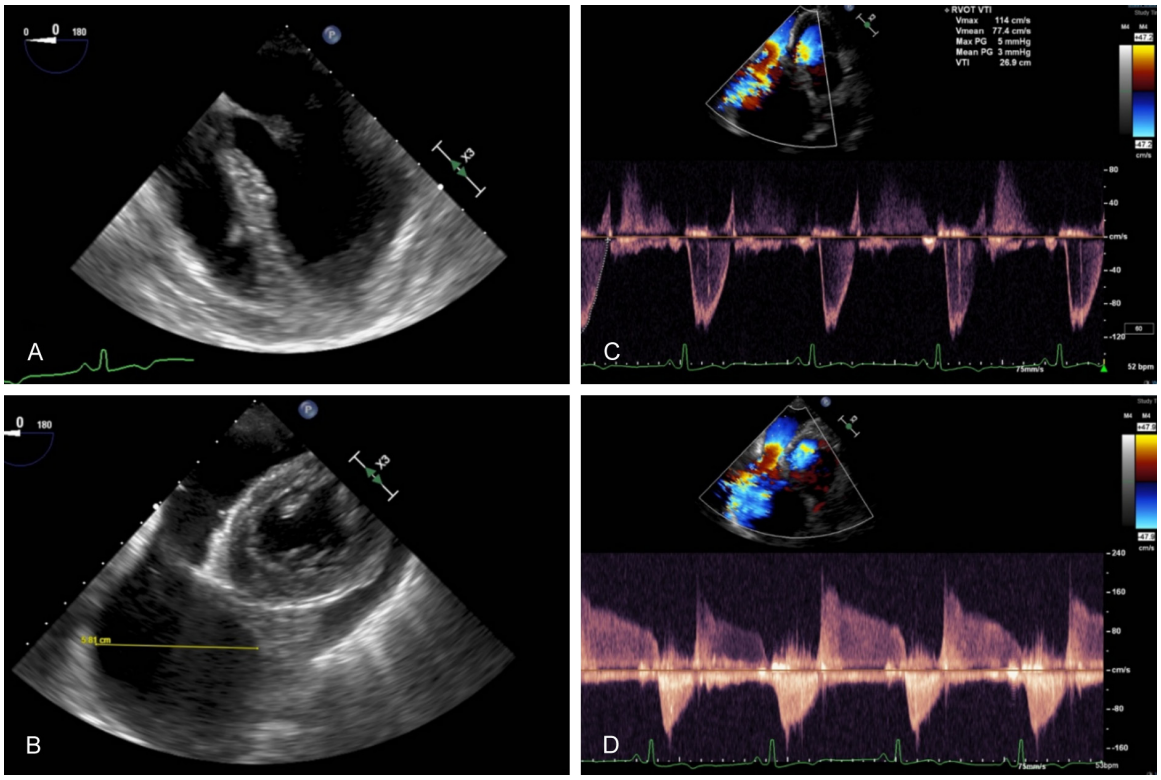


**Figure 2.** A: Transesophageal echocardiogram (TEE) showing normal left ventricular and right ventricular size. B: TEE deep gastric view showing aneurysmal main pulmonary artery measuring up to 5.8 cm in diameter. C: Deep gastric view of the TEE showing increased flow across right ventricular outflow tract with velocity time integral (VTI) of the RVOT 26.9 cm. D: Doppler study of the RVOT showing pulmonary regurgitation without evidence of pulmonic stenosis with trans-pulmonic maximal velocity (Vmax) 1.28 m/s.

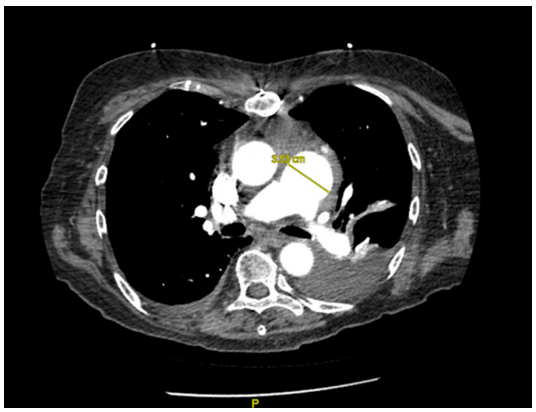

**Figure 3.** CTA of the chest after repair of the pulmonary artery using a 35-mm tubular graft.

monary emboli, iatrogenic, and malignancies of the lung, primary or metastatic [10]. Idiopathic PAAs are the least common etiology with the first reported case by Wessler and Jaches in 1923 [23]. In the literature review conducted by Gupta et al. [2], there were 254 cases of PAA reported in PubMed indexed publications between January 2010 and May 2018. Among the reported cases, 41 cases were identified as idiopathic with the mean age at presentation being 54 years and both genders equally affected. Additionally, 25 of the 41 cases were proximal with the average size 59 mm and 14 were distal aneurysms with the average size 38 mm [2].

As discussed earlier, currently, there are no clear guidelines for the management of PAAs [11]. The management should focus on treating the underlying conditions, periodic imaging follow-up and regular clinical evaluation [2]. The progression of PAA seems to be independent from pulmonary pressures during follow-up [24]. However, the duration of elevated PA pressure was an independent risk factor for the development of PAA [25]. Re-evaluation regularly for symptoms and with imaging should be taken into consideration for a change in treatment strategy if there is compression of adja-

cent structures, aneurysmal thrombus formation, increase in aneurysm diameter greater than 5 mm in 6 months, evidence of valvular abnormalities or shunt flow, and evidence of pulmonary arterial hypertension (PAH) [12, 20, 24].

Surgical management is reserved for symptomatic patients and PAA with high risk of aneurysmal rupture. The upper limit of PAA diameter deemed appropriate for surgical repair has been reported between 5-6 cm in different studies [10, 14, 18]. Surgical options include aneurysmorrhaphy, lobectomy, bilobectomy, aneurysmectomy, and pneumonectomy [2]. However, these techniques carry a high risk of morbidity and mortality, especially in those with a greater PAA diameter [10]. Additionally, patients with PAH should be considered for surgical management [10]. But, these patients with PAH have high surgical risk and may require transplantation [3, 10]. As in any case, the benefit of surgery should be weighed against the risk of surgery and underlying conditions should be taken into consideration in the decision-making process. Complications of surgery are composed of artery dissection, rupture, thrombosis, and airway compressions [18]. Despite that, due to the infrequent diagnosis and lack of published series of PAA patients, the true morbidity and mortality cannot be determined [10].

Endovascular treatment options such as coil embolization, stent-assisted coil embolization, and vascular plugs have been described for certain etiologies, particularly iatrogenic causes, saccular aneurysms, and small branches [3, 19]. Endovascular techniques have been reported to carry a much lower morbidity and mortality than surgical techniques [3, 18].

## Conclusion

PAAs remain a rare diagnosis with no specific symptoms to lead to diagnostic investigation. When PAAs are found, it is usually incidental on imaging studies. Contrast-enhanced CT has emerged as the confirmatory test to identify PAAs. However, other modalities such as echocardiography and MRI may assist with further characterization and discovery of underlying etiologies.

Once PAAs have been identified, the treatment choice is another challenge which must be determined. In general, patients who remain asymptomatic and PAA diameters which remain stable can follow a more conservative approach with medications. However, if the PAA diameter is greater than 5 cm and patients develop symptoms and/or PAH should undergo surgical interventions. Additional consideration for endovascular repair is another option for PAA characteristics. There are no large studies to confirm morbidity and mortality of this patient population and no guidelines for the management of these patients. Additional research needs to be conducted to clarify the definition and management of PAAs.

## Disclosure of conflict of interest

None.

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